

Conclusions: CVD is at least as common in this Djiboutian community as in other African cohorts of children. The absence of surgery was a major mortality risk factor. Dilated cardiomyopathy was frequent in this study. Much work remains to be done to discover the size and nature of genetic and environmental contributions to these various forms of pediatric heart diseases in the Horn of Africa.

340

Population-based evaluation of a suggested anatomic and clinical classification of congenital heart defects based on the International Paediatric and Congenital Cardiac Code

Lucile Houyel [Orateur] (1), Babak Khoshnood (2), Robert H. Anderson (3), Nathalie Lelong (2), Anne-Claire Thieulin (2), François Goffinet (2), Damien Bonnet (4)

(1) Centre Chirurgical Marie Lannelongue, M3C, Chirurgie des cardiopathies Congénitales, Le Plessis-Robinson, France – (2) Inserm UMR S953, UPMC, Université Paris-6, Paris, France – (3) Institute of Child Health, University College, Cardiac unit, London, Royaume-Uni – (4) AP-HP, CHU Necker-Enfants Malades, M3C, Cardiologie Pédiatrique, Paris, France

Background: Classification of the overall spectrum of congenital heart defects (CHD) has always been challenging, because of the diversity of the cardiac phenotypes and the often complex associations. The purpose of the study was to establish a comprehensive and easy-to-use classification of CHD for clinical and epidemiological studies based on the long list of the International Paediatric and Congenital Cardiac Code (IPCCC).

Methods: We coded each individual malformation using six-digit codes of the long list of IPCCC. We then regrouped all lesions into 10 categories and 22 subcategories according to a multi-dimensional approach encompassing anatomic, diagnostic and therapeutic criteria. This anatomic and clinical classification of congenital heart disease (ACC-CHD) was then applied to data acquired from a population-based study of CHD in France, including 2867 cases (82% live births, 1.8% stillbirths and 16.2% pregnancy terminations).

Results: The majority of cases (79.7%) could be identified with a single IPCCC code. The category “Isomerism and visceral heterotaxy” was the only one that typically required more than one code for identification of cases. The two largest categories were “ventricular septal defects” (52%) and “anomalies of the outflow tract and arterial valves” (20% of cases).

Conclusion: Our proposed classification is not new, but rather a regrouping of the known spectrum of CHD into a manageable number of categories based on anatomic and clinical criteria. The classification is designed to use the code numbers of the long list of IPCCC but can accommodate ICD-10 codes. Its exhaustiveness, simplicity, and anatomic basis make it useful for clinical and epidemiologic studies, including those aimed at assessment of risk factors and outcomes. The proposed classification can also provide a structure for various clinical and epidemiologic databases.

341

Parental electrocardiographic screening identifies a high degree of inheritance for congenital and childhood non-immune isolated atrio-ventricular block

Alban-Elouen Baruteau [Orateur] (1), Albin Behaghel (2), Swanny Fouchard (3), Christian Dina (3), Elisabeth Villain (4), Jean-Benoît Thambou (5), François Marçon (6), Véronique Gournay (7), Francis Rouault (8), Alain Chantepie (9), Sophie Guillaumont (10), François Godart (11), Caroline Bonnet (12), Alain Fraisse (13), Jean-Marc Schleich (2), Jean-René Lusson (14), Yves Dulac (15), Raphaël Martins (2), Stéphanie Chatel (3), Jean-Jacques Schott (3), Jean-Claude Daubert (2), Hervé Le Marec (16), Philippe Mabo (2), Vincent Probst (16)

(1) Centre Chirurgical Marie Lannelongue, Le Plessis Robinson, France – (2) CHU Pontchaillou, Cardiologie et Maladies Vasculaires, Rennes, France – (3) CHU Nantes, Institut du thorax, Inserm UMR 915 – CNRS ERL 3197, Nantes, France – (4) AP-HP, CHU Necker-Enfants-Malades, Cardiologie Pédiatrique, Paris, France – (5) CHU Bordeaux, Cardiologie pédiatrique, Bordeaux, France – (6) CHU Nancy, Cardiologie pédiatrique, Nancy, France – (7) CHU Nantes, Cardiologie Pédiatrique, Nantes, France – (8) Cabinet de Cardiologie Pédiatrique, Marseille, France – (9) CHU Tours, Cardiologie

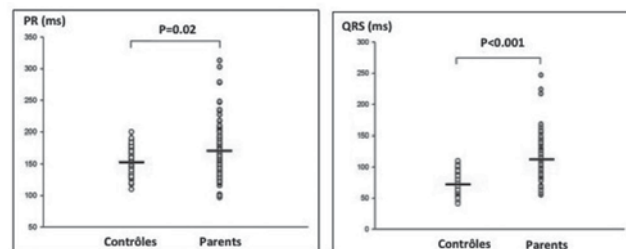
Pédiatrique, Tours, France – (10) CHU Montpellier, Cardiologie Pédiatrique, Montpellier, France – (11) CHU Lille, Cardiologie Pédiatrique, Lille, France – (12) CHU Dijon, Cardiologie Pédiatrique, Dijon, France – (13) CHU Marseille, Cardiologie Pédiatrique, Marseille, France – (14) CHU Clermont-Ferrand, Cardiologie Pédiatrique, Clermont-Ferrand, France – (15) CHU Toulouse, Cardiologie Pédiatrique, Toulouse, France – (16) CHU Nantes, Institut du Thorax, Cardiologie, Nantes, France

Introduction: The etiology of congenital or childhood non-immune, isolated AV block remains unknown. We hypothesized that this conduction abnormality in the young may be a heritable disease.

Method: A multicenter retrospective study (13 French referral centers, from 1980 to 2009) allowed inclusion of 141 children with AV block diagnosed *in utero*, at birth or before 15 years of age, without structural heart abnormalities and without maternal antibodies. Parents and matched controls were investigated for family history and for ECG screening.

Results: In parents, family history of sudden death or of progressive cardiac conduction defect was found in 1.4% and 11.1% respectively. Screening ECGs from 130 parents (mean age 42.0 ± 6.8 years, 57 couples) were compared to 130 matched healthy controls. All parents were asymptomatic and in sinus rhythm, except one with unknown complete AV block. Conduction abnormalities were more frequent in parents than in controls, respectively found in 50.8% versus 4.6% ($p < 0.001$). Long PR interval was found in 18.5% parents but never in controls ($p < 0.001$). Complete or incomplete right bundle branch block was observed in 39.2% parents and 1.5% controls ($p < 0.001$). Complete or incomplete left bundle branch block was found in 15.4% parents and 3.1% controls ($p < 0.001$). Heritability estimate for isolated conduction disturbances was very high, calculated at 91% (standard error=1.019, $p = 2.10^{-16}$).

Conclusion: ECG screening in asymptomatic parents from children affected by idiopathic AV block revealed a high prevalence of conduction abnormalities with prolongation of intra-atrial, AV and/or intra-ventricular conduction delay. Heritability estimate confirmed a high contribution of genetic factors. These results support the hypothesis of an inheritable trait in congenital and childhood non-immune, isolated AV blocks.



Comparison of PR interval and QRS complex duration

342

Assessment of systo-diastolic ventricular function using tissue Doppler imaging after successful repair of aortic coarctation

Sana Ouali [Orateur] (1), Sami Hammas (1), Helmi Ben Salem (1), Slim Kacem (1), Elyess Neffeti (2), Rim Gribaa (1), Fahmi Remedi (2), Essia Boughzela (2)

(1) Hôpital Sahloul, Cardiologie, Sousse, Tunisie – (2) Hôpital Sahloul, Sousse, Tunisie

Purpose: the aim of the study was to assess the ventricular systolic and diastolic function after the successful repair of aortic coarctation using tissue Doppler imaging (TDI).

Methods: The study group consisted of 28 patients (mean age 12 ± 4.2 years) after the aortic coarctation (AoC) repair. The TDI parameters and the conventional echocardiographic indices of the left and right ventricular systo-diastolic function were analyzed and compared with the results obtained from 22 healthy controls.

Results: Patients with repaired aortic coarctation had significantly decreased systolic (Sa) tissue Doppler velocities at the lateral mitral (11.8 vs 14.7 cm/s, $p = 0.001$), tricuspid (16 vs 18.7 cm/s, $p = 0.009$), and septal (10.2 vs 12 cm/s, $p = 0.058$) annuli compared with controls. The early and diastolic